Stomach "gyri" with Protein Losing Enteropathy: Report of a New Perpetrator

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Stomach “gyri” with Protein Losing Enteropathy: Report of a New Perpetrator

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Introduction:

Ménétrier disease is a rare, acquired disease characterized by enlarged gastric folds in the fundus and body with foveolar hyperplasia, cystic dilation of gastric glands , reduced numbers of parietal and chief cells with hypochlorhydria and protein losing enteropathy. We present a case of HIV as a new perpetrator for Ménétrier disease causing protein losing enteropathy.

Case Presentation

Chief Complaint:

A 41-year-old hispanic male with AIDS, chronic hepatitis C, and a history of ascending colitis with cecal ulcer, presented with 3-month history of unintentional weight loss of 20 pounds and diffuse pain in epigastrium and RUQ, with associated diarrhea, nausea, and vomiting.

Physical Examination:

Pertinent positives: Tenderness in epigastrium, and ascites with anasarca.

Investigations:

Laboratory data: Severe hypoalbuminemia (2.0gm/dL), hyponatremia (127 meq/dL), peripheral eosinophilia- 25%.

Ascitic fluid analysis: SAAG of <1.1.

Initial esophagogastroduodenoscopy (EGD) with superficial gastric biopsy: Findings consistent with eosinophilic gastritis.

Stool studies: Occasional Charcot-leyden crystals, ova and parasites negative.

CMV DNA: Negative.

Repeat EGD: Enlarged gastric fold in fundus and cardia with diffusely erythematous, nodular and friable mucosa.

Endoscopic ultrasound with full thickness gastric snare biopsy: Foveolar hyperplasia with superficial erosions associated with acute on chronic inflammation and sparse fundic glands with reactive changes (Fig. 1& 2).

Treatment:

Prednisone 40 mg + Octreotide 0.1 mg SQ BID for 5 days followed by monthly intramuscular injection.

Albendazole 400 mg for 3 days for possibility of Ancylostoma caninum (empiric).

Discussion:

Overexpression of transforming growth factor alpha has been proposed to play a role in the pathophysiology of Ménétrier’s disease. Its levels are increased in the gastric mucosa of patients with Ménétrier’s disease. TGF-alpha is responsible for maintaining gastric homeostasis by inhibiting acid secretion while increasing gastric mucous production. HIV-1 tat gene product stimulates TGF-alpha production, which activates the EGF receptor. Our hypothesis states that overexpression of TGF-alpha by HIV can lead to gastric epithelial changes consistent with Ménétrier’s disease via the pathway shown (Fig. 4).

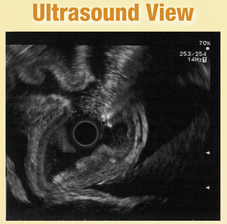

Figure 1

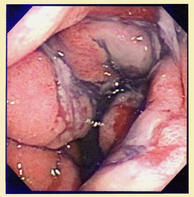

Figure 2

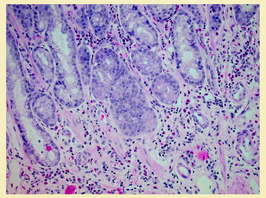

Figure 3

Our Hypothesis

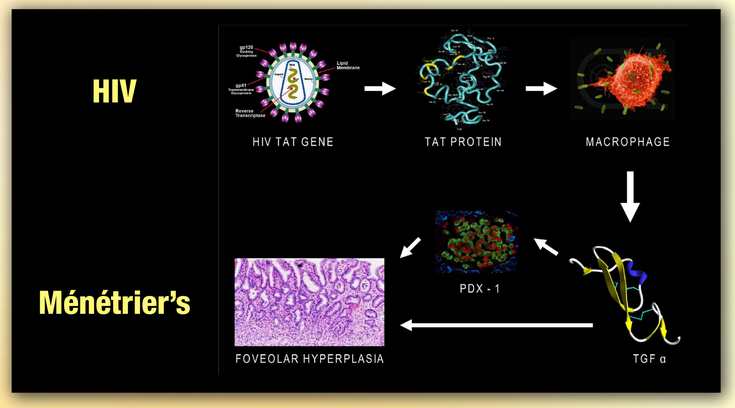

Figure 4. HIV induced upregulation of TGF-alpha, leading to Ménétrier disease. TGF: Tumor growth factor, PDX-1: Pancreatic and duodenal homeobox 1.

Ménétrier disease

(Other names include Hypoalbuminemic hyperplastic gastropathy and hyperplastic hypersecretory gastropathy)

Prevalence:

Very rare with fewer that 400 cases worldwide

Clinical Manifestations:

- Epigastric pain - 65 percent
- Asthenia - 60 percent
- Anorexia - 45 percent
- Weight loss - 45 percent
- Edema - 38 percent
- Vomiting - 38 percent

Diagnosis:

- Gross: Striking enlargement of gastric folds or rugae, confined to the body and fundus.
- Histology: Extreme foveolar hyperplasia with glandular atrophy on a full thickness biopsy (or endoscopic snare or suction biopsy).

Treatment:

- Octreotide has been shown to improve enteral protein loss. No clear data or study is available for duration of treatment.
- Case reports for treatment with a monoclonal antibody directed against the epidermal growth factor receptor (Erbitux).
- Surgical option with total gastrectomy.

Prognosis:

The natural history of Ménétrier’s disease is not well characterized. Scattered reports have demonstrated the evolution from Menetrier’s disease to gastric atrophy over four to eight years, with return of the serum albumin concentration to normal The risk of gastric cancer in patients with Ménétrier’s disease is uncertain, Other reports indicate that sepsis, vascular and/or thromboembolic complications may be a greater threat to patients with Menetrier’s disease than malignant transformation.

Key Points:

- Ménétrier disease is a rare cause of gastric rugal hypertrophy with protein losing enteropathy likely caused by elevated levels of transforming growth factor-alpha (TGF-α).
- AIDS patients may have higher risk of Ménétrier disease due to HIV-1 tat gene related over production of TGF-α.
- Ménétrier disease should be considered in an AIDS patient with abdominal pain and protein losing enteropathy who has no evidence of gastrointestinal CMV or H. pylori infection.